HHS Public Access

Author manuscript

Muscle Nerve. Author manuscript; available in PMC 2015 August 13.

Published in final edited form as:

Muscle Nerve. 2015 June ; 51(6): 815–821. doi:10.1002/mus.24488.

AMYOTROPHIC LATERAL SCLEROSIS SURVEILLANCE IN BALTIMORE AND PHILADELPHIA

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Abstract

Introduction—Limited epidemiological data on amyotrophic lateral sclerosis (ALS) exist in defined geographic areas in the United States.

Methods—Neurologists submitted case reports for patients under their care between January 1, 2009, and December 31, 2011, who met the El Escorial criteria. Diagnosis was confirmed for a sample of cases by the consulting neurologist. Death certificate data were used for supplemental case identification.

Results—The 248 reported cases were most likely to be 50–69 years old, men, white, and non-Hispanic. The total crude average annual incidence rate was 1.46 per 100,000 person-years.

Conclusions—The reported demographic characteristics were consistent with previously published findings. The crude annual incidence was slightly lower than the expected rate of 1.6 but was within the range reported previously (0.7–2.5). These findings help quantify the burden of ALS in the United States.

Keywords

amyotrophic lateral sclerosis; Baltimore; epidemiology; incidence; motor neuron disease; Philadelphia; surveillance

Amyotrophic Lateral Sclerosis (ALS) is a rare, progressive, and fatal neurological disease affecting both the upper and lower motor neurons. Familial forms of ALS account for 5–10% of cases.[1,2] The etiology of the majority of sporadic ALS cases is unknown, and several areas are under investigation, including environmental exposures, occupational exposures, physical activity, trauma, and genetic factors.[3–9] ALS is diagnosed based on the evaluation

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Disclaimer: The findings and conclusions in this report are those of the authors and do not necessarily represent the views of the Agency for Toxic Substances and Disease Registry.

Presentation of Material: Partial content found within this manuscript was presented as a poster at the joint annual meeting of the International Congress of Interventional Neurology and Pennsylvania Neurological Society, Philadelphia, PA, October 2013; and at the 12th Annual Northeast ALS (NEALS) Consortium Meeting, Clearwater, FL, October 2013.

of signs and symptoms and electromyogram (EMG) results. The El Escorial criteria use this information to classify the certainty of the diagnosis and are used primarily in research studies.[10]

Worldwide, the crude incidence rate of ALS is estimated to be 1.6 (range, 0.7 to 2.5) per 100,000 person-years, and ALS disproportionately affects those who are older, male, white, and non-His-panic.[11–18] The median duration of time from symptom onset to diagnosis has been reported to range from 10 to 36 months.[14,16] Much of the published epidemiological data regarding ALS are from outside of the United States (U.S.), and case ascertainment methodologies vary to include the use of existing data sets, neurologist reports, death certificates, hospital records, and pharmacological records.[11]

Uncertainty about incidence and prevalence in the United States supports the need for an ALS surveillance system, such as the congressionally mandated National ALS Registry (Registry), which was created and is maintained by the federal Agency for Toxic Substances and Disease Registry (ATSDR).[19] The Registry identifies ALS cases using national administrative datasets and patient self-registration through a web portal.[20] This nontraditional case ascertainment required validation through conventional surveillance techniques; therefore, ATSDR established state and metropolitan area-based surveillance projects.

The objectives of the surveillance projects were to obtain reliable and timely information regarding local incidence and demographic characteristics of ALS in defined geographic areas. This article summarizes the data collection methods used in the Baltimore and Philadelphia projects and describes the demographic characteristics of ALS in these areas.

MATERIALS AND METHODS

Project Sites

The Baltimore and Philadelphia projects are examined together due to their similarities. Both project areas are located in mid-Atlantic states and are within a 1-hr train ride along the Northeast Corridor.[21] Several large, academic medical institutions housing ALS referral centers that see upward of 50 or more ALS patients per year can be found in these 2 cities. Both areas have large black/African American populations compared with the entire U.S. population.[22] The median household income in 2010 for the City of Baltimore was $40,803, and for Philadelphia it was $37,016.[22] Other similarities include the number of individuals below the poverty line (23.4% in Baltimore and 26.2% in Philadelphia), the percentage with a high school degree or higher (79.6% in Baltimore and 80.4% in Philadelphia), and median age (34.4 years in Baltimore and 33.5 years in Philadelphia).[22] Finally, the data collection methodologies, including project staff, were similar for both projects.

For the Baltimore project, the catchment area included the City of Baltimore, Baltimore County, and Howard County, Maryland, and for the Philadelphia project, the catchment area included Philadelphia County, Pennsylvania, which is also known as the City of Philadelphia. According to the U.S. Census Bureau, American Community Survey, Baltimore had a population of 1,713,075 persons in 2010.[22] The population was 51.5%

white, 38.3% black/African American, 5.6% Asian, and 4.6% other, while 4.5% of the population was Hispanic.[22] Philadelphia had a population of 1,526,006 persons in 2010, and the breakdown by race and ethnicity was 41.0% white, 43.4% black/African American, 6.3% Asian, and 9.3% other, while 12.3% of the population was Hispanic.[22] This contrasts with a total U.S. population that was 72.4% white, 12.6% black/African American, 4.8% Asian, 10.2% other, and 16.3% Hispanic in the year 2010.[23]

Neurologist Identification and Recruitment

For both project sites, the American Medical Association list of neurologists' contact information was purchased from Medical Marketing Services, and additional information was added to the list from Internet-based searches and ALS Association recommendations. Neurologists specializing in the diagnosis/care of persons with ALS at ALS referral centers in Baltimore, MD, Philadelphia, PA, and Washington, DC, were identified for both projects, and special emphasis was placed on recruiting them to participate. Pediatric neurologists, neurosurgeons, and medical residents were excluded, as they are unlikely to see ALS patients. All remaining neurologists on the list were contacted by means of letters, phone calls, faxes, and face-to-face site visits to determine if they diagnosed or provided care to ALS patients. A neurologist could have received an introductory letter, as well as a combination of 7 phone calls and/or faxes, an office visit, and a "hard-to-reach" letter to determine if the neurologist diagnosed and/or provided care to ALS patients. Neurologists who did diagnose and/or provide care could have received a combination of up to an additional 7 phone calls and faxes, an office visit, and a "hard-to-reach" letter by means of Express mail to recruit them to report their cases.

Data Collection

Neurologists who diagnosed and/or provided care to ALS patients were asked to submit case reports for eligible patients. Eligible patients were under a doctor's care at any time between January 1, 2009, and December 31, 2011, met 1 of the El Escorial criteria (definite, probable, probable-laboratory supported, possible, not classifiable),[10] and had an address in 1 of the respective catchment areas at some point in the reporting period. Multiple reports for the same patient were accepted if they were reported by different neurology practices to assure complete case ascertainment and to obtain as much case information as possible.

The one-page Case Reporting Form included questions about patient demographic characteristics, month and year of symptom onset, month and year of diagnosis, and El Escorial criteria classification level. A Medical Record Verification Form and EMG report were requested on a sample of all case reports, before the de-duplication process, to allow a consulting neurologist specializing in the care/treatment of ALS to confirm ALS diagnosis.

Death certificates for decedents residing in the project's catchment areas were identified using the International Classification of Diseases-10 code for motor neuron disease (G12.2)[24] for the period 2009–2011. Attempts were made to obtain case reports for decedents for which the cause of death was explicitly stated as ALS who were not already reported. Compensation was offered for completed forms. No patients were contacted. The Baltimore and Philadelphia projects followed the Centers for Disease Control and

Prevention Institutional Review Board-approved State and Metropolitan Area-Based ALS Surveillance project protocol.

Data Cleaning and Analysis

All case reports were accepted, and duplicate cases were identified by matching on a combination of variables including last name, first name, address, date of birth, gender, and last 5 digits of the social security number. The process to create a composite, de-duplicated record for each case reported more than 1 time started with the case report containing the most complete information. The duplicate report, as well as the death certificate, when available, were used to fill in or clarify some demographic information. If there was conflicting information between the duplicate case reports and there was no death certificate, the response was changed to "unknown."

To calculate incidence rates, cases with a year of diagnosis in 2009, 2010, and 2011 were included in the analysis. Crude annual incidence rates were calculated for Baltimore, Philadelphia, and the 2 areas combined using the count of cases diagnosed in each year as the numerator and the corresponding 2010 U.S. Census population[22] data as the denominator. The crude average annual incidence rates were calculated by adding the incidence rates for each of the 3 years and then dividing by 3. Age-adjusted average annual incidence rates by gender and race were standardized to the year 2000 U.S. Standard Population.[25] The age-specific average incidence rates were calculated by dividing the count of all cases diagnosed in 2009 to 2011 in each age stratum by the corresponding 2010 U.S. Census population data and then dividing that number by 3. All incidence rates are listed as cases per 100,000 person-years.

All cases with a month and year of symptom onset and a month and year of diagnosis were included in the calculation of time from symptom onset to diagnosis. Time was calculated by subtracting the year of symptom onset from the year of diagnosis, multiplying by 12, and adding the difference between the month of diagnosis and the month of symptom onset. The continuous variable was then transformed into a categorical variable for presentation in this report. Data were analyzed using Microsoft Excel®[26] and SPSS, version 19.0.[27]

RESULTS

Reporting Providers

All 608 neurologists were contacted in the combined project area; 7.6% (46/608) diagnosed and/or provided care to ALS patients in the project's time period; and 32.6% (15/46) of them reported eligible cases. An additional 17.4% (106/608) of neurologists would diagnose and/or care for ALS patients but did not have prevalent or incident cases for the period 2009–2001 to report. Overall, 75.0% (456/608) of neurologists said they would not diagnose and/or care for ALS patients, including 259 neurologists affiliated with academic medical centers.

Reporting Practices

The 608 neurologists represented 214 practices. Neurologists at 8.9% (19/214) of the practices diagnosed and/or provided care to ALS patients in the project's time period, and neurologists at 63.2% (12/19) of them reported cases. Neurologists at 23.8% (51/214) of the practices would diagnose and/or care for ALS patients but did not have prevalent or incident cases for the period 2009 to 2011 to report. Neurologists at the remaining 67.3% (144/214) of practices would not diagnose and/or care for ALS patients.

Validation of Case Reports

Before de-duplicating the case dataset, 27 Medical Record Verification Forms, representing 10.1% (27/267) of collected case reports, were requested, and 25 were received. The consulting neurologist determined that 72% (18/25) of these cases were "definite", "probable", or "probable laboratory-supported" ALS as per the El Escorial criteria. The remaining 28% (7/25) were determined to be "possible" ALS based on the El Escorial criteria.[10] The reporting providers and consulting neurologist agreed that all cases did have ALS. The reporting providers and consulting neurologist classified 52% (13/25) of the cases exactly the same, and 96% (24/25) of the cases were classified as either "definite," "probable," "probable-laboratory supported," and "possible."

Case Ascertainment

Using 2010 U.S. Census population statistics and estimates of incidence and prevalence, we expected 253 unique cases in the combined project area.[11,22] A total of 267 case reports were received, and after de-duplication there were 248 unique cases in the de-duplicated dataset, which is approximately 98% (248/253) of the expected cases. Ninety percent of cases reported were submitted by ALS specialists at ALS referral centers in the areas, and 10% of cases reported were submitted by neurologists from general neurology practices.

El Escorial Criteria Classification of ALS Cases

In the combined project area, 88.7% (220/248) of cases were reported as "definite," "probable," or "probable-laboratory supported" ALS; 10.5% (26/248) were reported as "possible" ALS; and 0.8% (2/248) were reported as "not classifiable" according to the El Escorial criteria. For the 2 cases reported as "not classifiable" by the reporting provider, 1 of these cases were verified as "probable" by the consulting neurologist and the other was matched to a death certificate that explicitly stated ALS on it.

Demographic Distribution of Cases

In Baltimore, Philadelphia, and the 2 sites combined, reported cases were most likely to be men, white, and non-Hispanic (Table 1). In Baltimore, the percentage of reported cases in each age group increased until ages 60–69, at which time the percentage began to decrease. A similar trend occurred in Philadelphia until the 50–59 age group. The Philadelphia patients were slightly younger than the Baltimore patients at age of diagnosis. In Philadelphia, 31.6% (36/114) of the reported patients were black/African American, while in Baltimore, 21.6% (29/134) were black/African American. In Baltimore, no Hispanic patients were reported, and in Philadelphia 5 Hispanic patients were reported.

Incidence

Sixty-two percent (83/134) and 62.8% (59/114) of cases were diagnosed in 2009, 2010, and 2011 in Baltimore and Philadelphia, respectively, for a total of 142 incident cases in the combined project area. Combined, the crude annual incidence rates for 2009, 2010, and 2011 were 1.51, 1.54, and 1.33 per 100,000 person-years, respectively, and the average crude annual incidence rate was 1.46 per 100,000 person-years. The crude average incidence rate was somewhat higher in Baltimore compared with Philadelphia (1.61 vs. 1.29 per 100,000 person-years). Age-specific average annual incidence rates trended upward with age, then decreased in Philadelphia and the combined project area for the oldest age group (Table 2). The total age-adjusted average annual incidence rate was 1.40 per 100,000 person-years for the combined project area. There was a difference between age-adjusted rates in men (1.74 per 100,000 person-years; 95% confidence interval [CI], 1.34–2.14) and women (1.13 per 100,000 person-years; 95% CI, 0.85–1.41), however this difference did not differ statistically (Table 3). The age-adjusted rates between whites-only and blacks/African Americans-only in Philadelphia and the combined project area differed statistically (Table 3).

Time from Symptom Onset to Diagnosis

Twenty-two cases were removed because they were missing data. Fifty percent of cases in the combined project area had symptoms for less than 12 months before diagnosis; 90% had symptoms for up to 32 and 36 months before the date of diagnosis in Baltimore and Philadelphia, respectively.

DISCUSSION

Due to the severity of motor neuron diseases, all patients should be seen by a neurologist at least 1 time during the course of their disease.[28] To date, few epidemiological studies of ALS incidence have been conducted in the United States using a case ascertainment methodology designed to collect case reports for all ALS patients from all neurologists who diagnose and/or care for ALS patients in a defined geographic area. Rather, most studies use clinical samples or death certificate data to generate incidence rates. Furthermore, an understanding of the proportion of neurologists who diagnose and/or care for ALS patients, accurate estimates of the incidence of ALS, and a demographic characterization of people affected by ALS in the United States are not readily available. This information could be useful to health agencies, health care providers, patient services groups, and public health practitioners. This report provides a summary of data collection methods and epidemiological information about ALS cases in the metropolitan areas of Baltimore, MD and Philadelphia, PA.

All neurologists in the Baltimore and Philadelphia areas were contacted and encouraged to participate in order to have unbiased case ascertainment. Nearly 80% of all neurologists, representing 67% of all practices, did not diagnose and/or provide care for persons with ALS. Much of this can be attributed to the large proportion of neurologists who are affiliated with the academic medical centers in these 2 metropolitan areas; specifically, 47.5% and 37.2% of the total number of neurologists in Baltimore and Philadelphia, respectively.

However, at least 1 neurologist specializing in the diagnosis/treatment of persons with ALS at all ALS referral centers housed at the academic medical centers in Philadelphia, PA, Baltimore, MD, and Washington, DC participated, and 90% of all case reports were submitted by these centers.

Attempts were made to obtain case reports for decedents for whom the cause of death was explicitly stated as ALS on their death certificates. These efforts yielded an additional 24 case reports (17.9% of reported cases) in Baltimore and 13 (11.4% of reported cases) in Philadelphia. The additional case reports were crucial for describing the demographic characteristics of ALS patients and calculating incidence rates in these 2 metropolitan areas. Future surveillance projects should consider the use of death data as a supplemental tool to ascertain cases.

Despite our efforts, including offering compensation for completed forms, we were unsuccessful in gaining the participation of 7 practices, 2 in Philadelphia and 5 in Baltimore. The Baltimore Veterans Affairs Medical Centers (VAMC) submitted case reports for eligible decedents only, and the Philadelphia VAMC did not participate. Furthermore, a review of the death certificate data yielded 23 decedents in Baltimore and 9 decedents in Philadelphia in whom ALS was listed as a cause of death that were not reported. It is uncertain how many of these decedents would have been reported as actual ALS cases by a neurologist. It is likely that some cases went unreported in both project areas, but we are unable to determine exactly how many. However, based on current epidemiological estimates, we are confident that a very large proportion of eligible cases were reported to the project.

As the literature indicates, reported cases in Baltimore and Philadelphia were more likely to be older, men, white, and non-Hispanic.[11–18] Overall, the demographic characterization of ALS cases is similar to reported findings of a surveillance project using similar methods and conducted on a state-wide scale in New Jersey.[18] The results we found regarding the time between symptom onset and diagnosis also agree with previous research.[14,16]

We found the combined crude annual incidence rates for 2009, 2010, and 2011 to be 1.51, 1.54, and 1.33 per 100,000 person-years, which were slightly lower than the expected rate of 1.6 per 100,000 person-years, but within the range (0.7–2.5) reported previously.[11] If the 32 decedents identified were true ALS cases and had been reported, the incidence rates would still be within the currently published range. Another possible explanation for the lower than expected incidence rates is that the percentage of minorities (specifically blacks/African Americans) in Baltimore and Philadelphia is larger than the U.S. population, and studies have shown that blacks/African Americans have a lower incidence rate for ALS than whites.[18,29,30]

The data suggest a difference in age-adjusted average annual incidence rates among men (1.74 per 100,000 person-years; 95% CI, 1.34–2.14) compared with women (1.13 per 100,000 person-years; 95% CI, 0.85–1.41), which is consistent with previously reported findings in the United States.[11,13,18] Furthermore, the data demonstrate a statistical difference in age-adjusted average annual incidence rates among whites-only (1.90 per

100,000 person-years; CI, 1.52–2.27) compared with blacks/African Americans-only (0.85 per 100,000 person-years; CI, 0.55–1.16), which is also consistent with the literature.[18,29,30] We were unable to calculate age-adjusted average annual incidence rates by all racial and ethnic groups due to the small number of cases in the subgroups. It is possible that if we had conducted surveillance over a longer period of time, and therefore had more cases, we may have had more power to detect differences. A paucity of studies exist that examine ALS incidence rates by race and ethnicity. Future research should be conducted to identify differences in incidence rates by race and ethnicity in the United States.

Conducting surveillance for a nonreportable chronic condition was challenging, time-consuming, costly, and would be difficult to implement on a national basis. It has been suggested the ATSDR evaluate the completeness of the National ALS Registry.[31] These findings, along with findings from the other state and metropolitan area-based surveillance projects, help to more accurately quantify the burden of ALS in the United States and will help to validate data in the National ALS Registry. This project has demonstrated the importance of well-developed case ascertainment strategies to populate the National ALS Registry and may aid with future research endeavors.

Acknowledgments

The authors thank the state and local health departments and other organizations that assisted with data collection, Dr. Eric Sorenson from the Mayo Clinic for serving as a consulting neurologist, and Dr. Jerald Fagliano, Program Manager, Environmental and Occupational Health Surveillance Program, New Jersey Department of Health for his technical expertise. This project was funded by the Agency for Toxic Substances and Disease Registry (Contract #200-2010-F-36614).

Abbreviations

ALS	amyotrophic lateral sclerosis
ATSDR	Agency for Toxic Substances and Disease Registry
CI	confidence interval
EMG	electromyogram
Registry	National ALS Registry
U.S	United States
VAMC	Veterans Affairs Medical Centers

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Table 1

Demographic characteristics of all reported ALS cases for the period January 1, 2009, to December 31, 2011 in Baltimore, Philadelphia, and the combined project area, $n = 248$.

	Baltimore		Philadelphia		Combined	
	Count of cases	**%**	**Count of cases**	**%**	**Count of cases**	**%**
Age						
Under 30 years	3	2.2	5	4.4	8	3.2
30–39 years	6	4.5	6	5.3	12	4.8
40–49 years	12	9.0	17	14.9	29	11.7
50–59 years	29	21.6	29	25.4	58	23.4
60–69 years	43	32.1	26	22.8	69	27.8
70–79 years	23	17.2	24	21.0	47	19.0
80 years or older	16	11.9	6	5.3	22	8.9
Unknown	2	1.5	1	0.9	3	1.2
Gender						
Men	73	54.5	63	55.3	136	54.8
Women	61	45.5	51	44.7	112	45.2
Race						
Asian	2	1.5	4	3.5	6	2.4
Black/African American	29	21.6	36	31.6	65	26.2
White	99	73.9	70	61.4	169	68.2
Unknown	4	3.0	4	3.5	8	3.2
Other	0	0.0	0	0.0	0	0.0
Ethnicity						
Hispanic or Latino	0	0.0	5	4.4	5	2.0
Non-Hispanic or Latino	130	97.0	104	91.2	234	94.4
Unknown	4	3.0	5	4.4	9	3.6
Total	134	100.0	114	100.0	248	100.0

Table 2

Age-specific average annual incidence rates for ALS cases diagnosed in a 3-year period 2009 to 2011 in Baltimore, Philadelphia, and the combined project area, n = 142.

	Baltimore (*n* = 83)		Philadelphia (*n* = 59)		Combined (*n* = 142)	
	Count of cases	**Age-specific rate***	**Count of cases**	**Age-specific rate***	**Count of cases**	**Age-specific rate***
Age						
Under 30 years	2	0.10	2	0.10	4	0.10
30–39 years	2	0.30	2	0.33	4	0.31
40–49 years	3	0.41	11	1.91	14	1.07
50–59 years	19	2.63	16	2.84	35	2.73
60–69 years	27	5.68	14	3.69	41	4.80
70–79 years	16	6.16	11	4.81	27	5.53
80 years or older	14	6.91	3	1.79	17	4.60

*Each rate was calculated by dividing the count of cases in each stratum by the 2010 U.S. Census population data for that stratum[22], then dividing by 3 and is listed per 100,000 person-years.

Table 3

Age-adjusted average annual incidence rates by gender and race for ALS cases diagnosed in a 3-year period 2009 to 2011 in Baltimore, Philadelphia, and the combined project area.

	Baltimore			Philadelphia			Combined		
Demographic characteristic	**Count of cases**	**Age-adjusted rate***	**CI†**	**Count of cases**	**Age-adjusted rate***	**CI†**	**Count of cases**	**Age-adjusted rate***	**CI†**
Gender									
Men	44	1.83	1.28–2.39	32	1.63	1.05–2.20	76	1.74	1.34–2.14
Women	39	1.20	0.82–1.58	27	1.07	0.66–1.47	66	1.13	0.85–1.41
Race‡									
White only	65	1.88	1.41–2.35	40	1.96	1.33–2.59	105	1.90	1.52–2.27
Black/African American only	16	0.95	0.48–1.43	15	0.79	0.39–1.20	31	0.85	0.55–1.16
Total	83	1.48	1.15–1.80	59	1.31	0.97–1.65	142	1.40	1.16–1.63

*Each rate was age-adjusted to the 2010 U.S. Census data[22] and standardized to the year 2000 U.S. Standard Population[25] and is listed per 100,000 person-years.

†95% Confidence intervals.

‡In Baltimore, 1 patient with “Asian” race and 1 patient with “unknown” race were excluded. In Philadelphia, 2 patients with “Asian” race and 2 patients with “unknown” race were excluded.